HEREDITARY ANGIOEDEMA: UNDERSTANDING THE CONDITION, TREATMENT OPTIONS AND COSTS

AXIS | AXIS Re

Today's healthcare landscape is changing rapidly with new possibilities and challenges coming from every aspect of medicine, science and technology. While the cost of healthcare has skyrocketed in recent years, the specialty pharmaceutical area accounts for most of the total healthcare spend in the United States.

What were once rare diseases are being diagnosed more frequently with advances in technology. Add to that the advent of "fast track" Food and Drug Administration (FDA) approval for treatments of rare diseases. That makes an attractive and growing market for the pharmaceutical industry's specialty drugs. Prescription drug spending on specialty drugs is the fastest growing area of cost.[1]

Payers need to be familiar with these potential high-cost diagnoses and their treatments to avoid runaway claims. The costs accumulate so quickly. HAE is just one of those conditions.

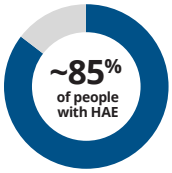
What is Hereditary Angioedema (HAE)?

Hereditary Angioedema (HAE) is a very rare genetic defect condition where individuals experience recurring episodes of severe swelling in multiple areas of the body such as the limbs, face, abdomen and airway. Individuals with HAE either have a low level of an important protein C1 esterase inhibitor (C1-INH) in their blood or the C1-INH protein does not function properly.[2]

There are three types of HAE[3,4]

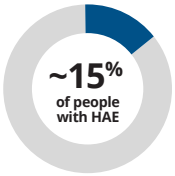
TYPE I

This is the most common form where people do not make enough protein C1-INH. This leads to painful swelling.



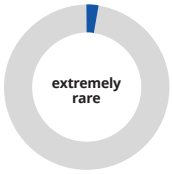
TYPE II

These people have normal or higher levels of C1-INH, but the protein doesn't work as it should, which can cause debilitating pain and may be life-threatening.



Type III or HAE with normal functioning C1-INH

This occurs mostly in women, and it is very rare and not well understood.



Why should you know about HAE?

There is no cure for HAE. Currently, 97% of the treatment costs for HAE are medications.[5] In addition to the high cost of targeted medications, patients can incur other costs related to relief of the symptoms of swelling and pain.

Potential HAE treatment costs

(per year)

$2 million+
in prescription drug charges per year*
*2018 AXIS Re Claims experience

$3–$5 million
for acute and prophylactic pharma therapy per year*
*AXIS partners and service providers

$600,000–$1 million
annually in lifetime maintenance costs*
*Average Wholesale Price (AWP) derived from Redbook

HAE treatment and related costs

There are two types of HAE-related treatment costs: 1) the cost of acute flare-ups, and 2) the cost of ongoing treatment with preventative medications.

The current HAE treatment includes seven Food and Drug Administration (FDA)-approved medications that are grouped into those two categories: 1) prophylaxis preventing an HAE attack, and 2) treatment for an acute HAE episode. It's important to know which drugs are approved for which use.

HAE Annual Drug Costs[6]

Routine preventative use

Specialty drug	Conservative estimated annual cost	Severe estimated annual cost
Cinryze®	$584,948	$777,788
Haegarda®	$615,888	$818,928
Takhzyro®	$635,616	$1,271,232

Acute episode treatment

Specialty drug	Conservative estimated annual cost	Severe estimated annual cost
Berinert® (acute)	$63,828	$255,312
Firazyr® (acute)	$240,785	$963,143
Kalbitor®	$318,978	$1,275,912
Ruconest® (acute)	$82,200	$328,800

HAE Claims Scenarios

HAE attacks are unpredictable, greatly differ from person to person, and require a lifelong treatment plan. Because no treatment is the same among patients, management of HAE is crucial to control costs and prevent further complications.

To help you understand the potentially significant financial impact of the high cost medications used to treat HAE, we have provided case studies outlining estimated annual cost based on Average Wholesale Price (AWP).

Robert

severe case with multiple acute episodes

Robert was taking three drugs for HAE: Haegarda, Ruconest and Firazyr

Haegarda is indicated for routine prophylaxis to prevent attacks. There was a shortage of this drug in early 2018. Because this preventative drug was unavailable, it increased Robert's acute attacks to twice a month. The other two drugs, Ruconest and Firazyr, are indicated for use in acute attacks.

$531,800
Firazyr
payment total

$1,689,679
Ruconest
payment total

$2,221,479[7]
Total Rx spend
on HAE drugs per year

Amy

severe case with multiple acute episodes

Amy was receiving Cinryze for routine preventative treatment. However, treatment changed to a different drug therapy during the Plan year. The revised medication protocol included Ruconest for prophylaxis and Firazyr for treatment of acute episodes.

While Firazyr is FDA-approved for treatment of acute attacks of HAE, Ruconest is not approved for prophylactic use. The combination of Firazyr and Ruconest is not FDA-approved and is considered an off-label use. Ruconest for prophylactic use of HAE is in a Phase II Clinical Trial. As the Plan document excluded the use of drugs in a clinical trial, these charges were ineligible.

$334,835
Firazyr
payment total

$1,648,994
Ruconest
payment total

$1,983,829[7]
Total Rx spend
on HAE drugs per year

How can AXIS Re help?

AXIS Re understands the complexity of the healthcare claims environment. The goal is for quality improvements, better patient outcomes, and getting a pulse on loss exposures. When there is a rare medical condition tied to high claims costs, it can be difficult to navigate through all the mountains of information and identify the best practices for optimal care. We take on these challenges to help ensure clients receive cost-effective, quality care.

For more information on HAE or any other rare condition, please reach out to your contact at AXIS Re or email **AccidentReClaimsNAM@axiscapital.com**.

Download the full white paper

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References

1 **https://www.actuary.org/content/prescription-drug-spending-us-health-care-system**

2 **https://www.medicinenet.com/hereditary_angioedema_hae/article.htm**

3 **https://www.discoverhae.com/hereditary-angioedema-symptoms and https://ghr.nlm.nih.gov/condition/hereditary-angioedema**

4 **https://www.cslbehring.com/patients/find-your-disease/hereditary-angioedema**

5 **https://www.haea.org/pages/p/ApprovedTreatments**

6 Average Wholesale Price (AWP) derived from Redbook. For acute medications, conservative estimates are based on an average of six flare-ups per year, and severe estimates are based on 24 flare-ups per year.

All of these drugs may be administered at home after training, except for Kalbitor. This drug must be injected by a healthcare professional.

There are also additional costs related to medications to treat symptoms such as nausea and vomiting, severe pain and diuretics for the inflammation.

7 Case studies are based on AXIS Re and industry partner experience and are provided for illustrative purposes only. Examples may be based on actual cases, composites of actual cases or hypothetical claim scenarios. Facts have been changed to protect the confidentiality of the parties. Whether or to what extent a particular loss is covered depends on the facts and circumstances of the loss, the terms and conditions of the policy as issued, and applicable law.